Pulmonary hypertension – an update

Ulrich, Silvia

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Silvia Ulrich

Clinic for Pulmonology, University Hospital Zürich, Switzerland

Since the first description of pulmonary vascular disorders by Ernst von Romberg in 1891, decades of research have led to progresses in the pathogenetic understanding, clinical classification and therapy of pulmonary hypertension (PH) [1]. Milestones of the early PH-research epoch were the introduction of right heart catheterisation by Werner Forssmann 1926, the first World Health Organization (WHO) meeting on PH in Geneva 1973 (where PH was classified into primary and secondary, with primary PH distinguishing between "arterial-plexiform", "veno-occlusive" and "thromboembolic" forms) and the first randomised controlled trial showing the efficacy of intravenous prostanoids in the treatment of "primary" PH [2]. Now, more than a dozen different medical therapies are available to treat pulmonary arterial hypertension (PAH) [3], a surgical technique has been developed, which potentially cures chronic thromboembolic PH (CTEPH) [4] and survival has improved according to registries [5, 6]. We are looking back on five WHO meetings on pulmonary vascular disorders, with the most recent one held in Nice 2013, which have summarised the pathogenetic understanding, have continuously updated the classification according to current knowledge and have provided comprehensive overviews of therapies, resulting in treatment guidelines [3, 7–10]. This edition of «Cardiovascular Medicine» is meant to provide a comprehensive overview for cardiologists, internists and other specialists. The special issue covers the most important aspects of the pathogenesis, classification and treatment of PH, with special emphasis on the current medical treatment options for PAH group I and the management of CTEPH. A special article is dedicated to the most common forms of PH, namely PH associated with left heart disease and with chronic lung disease. Targeted therapies for the pulmonary vascular aspect of these diseases are still missing. As exertional dyspnoea and fatigue, the early PH symptoms, gradually develop, many patients may delay seeking help for months or even years, and caregivers often do not consider PH until the disease is progressed and investigations reveal signs of right heart failure. Thus, we hope that with these review articles we can sensitise practising cardiologists and other physicians to the PH disease spectrum in order to get the disease diagnosed as early as possible and patients referred to specialist centres where they will be classified and treated according to highest standards and enrolled into clinical trials to further advance the PH field.

References

1 Von Romberg E. [Über Sklerose der Lungenarterie] On sclerosis of pulmonary arteries. Dtsch Arch Klin Med. 1891–1892;48:197–206. (German)
2 Rubin LJ, Mendoza J, Hood M, McGoon M, Barst R, Williams WB, et al. Treatment of primary pulmonary hypertension with continuous intravenous prostacyclin (epoprostenol). Results of a randomized trial. Ann Intern Med. 1990 Apr 1;112(7):485–91. PubMed PMID: 2107780. Epub 1990/04/01.
3 Galie N, Hoeper MM, Humbert M, Torbicki A, Vachiery JL, Barbera JA, et al. Guidelines for the diagnosis and treatment of pulmonary hypertension. The task force for the diagnosis and treatment of pulmonary hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS), endorsed by the International Society of Heart and Lung Transplantation (ISHLT). Eur Respir J. 2009 Sep 24;34:1219–63. PubMed PMID: 19749199. Epub 2009/09/15.
4 Kim NH, Delcroix M, Jenkins DP, Channick R, Dartevelle P, Jansa P, et al. Chronic thromboembolic pulmonary hypertension. J Am Coll Cardiol. 2013 Dec 24;62(25 Suppl):D92–9. PubMed PMID: 24355646.
5 Fischler M, Speich R, Dorschner L, Nicod L, Domenighetti G, Tamm M, et al. Pulmonary hypertension in Switzerland: treatment and clinical course. Swiss Med Wkly. 2008 Jun 28;138(25–26):371–8. PubMed PMID: 18587689.
6 Benza RL, Miller DP, Barst RJ, Badesch DB, Frost AE, McGoon MD. An evaluation of long-term survival from time of diagnosis in pulmonary arterial hypertension from the REVEAL Registry. Chest. 2012 Aug; 142(2):448–56. PubMed PMID: 22281797.
7 Rich S. Executive summary from the World Symposium on Primary Pulmonary Hypertension, Evian, France, September 6–10, 1998, co-sponsored by The World Health Organization. 1999.
8 World symposium on pulmonary hypertension (Venice 2003). 2003.
9 Galie N, Simonneau G. The fifth world symposium on pulmonary hypertension. J Am Coll Cardiol. 2013 Dec 24;62(25 Suppl):D1–3. PubMed PMID: 24355633.
10 Simonneau G, Gatzoulis MA, Adatia I, Celermajer D, Denton C, Ghofrani A, et al. Updated clinical classification of pulmonary hypertension. J Am Coll Cardiol. 2013 Dec 24;62(25 Suppl):D34–41. PubMed PMID: 24355639.

Correspondence:
Silvia Ulrich, MD
Clinic for Pulmonology, University Hospital Zürich
Rämistr. 100
CH-8091 Zürich
Switzerland
silvia.ulrich[at]usz.ch